Giant congenital melanocytic naevi: definition, malignant transformation and treatment modalities
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Results of early use of tissue expansion for giant congenital melanocytic naevi of the face and scalp with a follow-up of nine years

Zaal LH
Van der Horst CM

Abstract

Introduction. Giant congenital melanocytic naevi (GCMN) are uncommon, have a significant morbidity and require extensive treatment. This paper presents results after complete excision of GCMN at the scalp, forehead or periorbita after early tissue expansion. Based on 15 years of experience, we want to show that performing tissue expansion at a young age is advisable.

Patients & Methods. We included 17 consecutive patients in whom 38 tissue expanders were used. Early and late complications were noted. Patients were seen for a clinical follow-up in which scars and re-pigmentation were evaluated with a validated scar scale (POSAS).

Results. All GCMN could be excised completely with early tissue expansion. The age at treatment ranged from four months to two years of age. With a mean follow-up period of 8.7 years, in only three patients mild re-pigmentation was seen and none of the patients developed a malignant melanoma. Complication rates are comparable with literature.

Conclusion. Tissue expansion is a good method for removing GCMN located at the scalp or face with good cosmetic and oncologic results. Performing tissue expansion at a young age is advisable.
Introduction

Giant congenital melanocytic naevi (GCMN) occur in 1:20,000-50,000 newborns.1 GCMN are disfiguring lesions that differ in clinical and histological appearances and may be associated with the development of malignancy, particularly malignant melanoma.1,2,3 Treatment has two goals: an improved cosmetic appearance and diminution of the risk of malignancy. Techniques advocated to treat GCMN are curettage, laser or excision. Since in curettage and laser only the superficial layer of the naevus cells can be removed, (early) prophylactic excision has gained more acceptance, but poses a major reconstructive challenge due to the extension of the GCMN.

Antonyshyn stated that tissue expansion (TE) is the treatment modality of choice for reconstruction of many scalp lesions.4 Tissue expansion can be used very early in infancy and provides real advantages: skin flaps with subcutaneous tissue, minimal blood loss, wound closure without tension, and a large excision after each expansion procedure.4 The purpose of this study was to evaluate the long term cosmetic and oncological results of excision and subsequent reconstruction in cases of GCMN on scalp and periorbita after early TE and the prevalence of major and minor complications, since these are seldomly described in literature.

Methods & Materials

CMN are defined as giant CMN (GCMN) whenever their size is larger than one percent of the Total Body Surface Area (TBSA) in the face/neck or more than two percent elsewhere on the body.5 As in burn wound measurements, the patient’s palm of the hand is taken as a reference for measuring one percent.

In this cross-sectional observational study all patients were included with GCMN involving scalp or periorbita, who needed TE as part of their operative management. If serial excision could be performed in two operations, patients were not scheduled for TE. Patients were only included with a minimum of one year after reconstruction. In the same period starting in 1991 93 patients with GCMN covering one to 30 percent TBSA were treated in our department. All patients were photographed pre- and postoperatively by a medical photographer. All excised naevi were submitted for histopathologic evaluation.

The patients were invited to our outpatient department for clinical follow-up of oncologic and cosmetic outcomes. After informed consent was obtained, scars were evaluated using a validated scare scale (POSAS) and parents and children were given a questionnaire.6
Scar assessment

The POSAS consists of two scales; the observer scale and the patient scale. If at time of follow-up the patient was eight years or younger, the parents scored the patient scale. Both scales contain six items that are scored numerically. Each of the six items on both scales has a 10-step score, with 10 indicating the worst imaginable scar or sensation. The total score of both scales consists of adding the scores of each of the six items (range 6 to 60). The lowest score, 6, reflects normal skin, whereas the highest score, 60, reflects the worst imaginable scar. The two observers (L.Z. and C.vdH.) rated scar vascularity, pigmentation, thickness, relief, and surface area. Because pigmentation is an important outcome in this study, this item is further specified in Table I. For the cosmetic outcomes, and if relevant, patterns of hair growth or brow asymmetry were described, skull asymmetry was noted. Parents were also asked if they are satisfied with the cosmetic results.

Surgical technique

The minimum age at treatment with TE at the scalp or face was four months, since the expander becomes heavier after infiltration and a more developed neck musculature is needed. In general crescent shaped but occasionally round expanders with separate subcutaneous inflation reservoirs were placed in the subgaleal plane. Antibiotic coverage was given pre-operatively and continued for 24 hours. Expansion started two weeks after implantation. The maximum amount of filling with physiologic salt for each patient, depended on the size of the expander, the tension of the skin during the filling procedure and the tolerance of the patient. Seldom anaesthetic ointment was used. The final volume was always the maximum volume of the expander. In 17 patients 38 expanders were used, with an average of 2.2 expanders per patient. In average 3.2 operations per patient were needed, in which 30 flaps at the scalp or face were used for reconstruction. The time between two consecutive expansions was two to five months, with a mean of two months. In the majority of the cases 250 cc expanders were used, the smallest was 70 cc, the largest 750 cc of volume.

Results

In our study GCMN at the scalp, forehead or face were excised completely after TE in 17 patients. All patients were seen for follow-up. Distribution of age, sex and location of GCMN are shown in Table 1. Patients were seen for follow-up after an average of 8.7 years (range 1.5 – 16 years). Table 1 shows the re-pigmentation and POSAS scores
Table 1. Patient characteristics, location of GCMN, POSAS score, re-pigmentation, cosmetic outcome

POSAS scores: the first number of the observer and patient score represents the total amount of adding the six separate items each with a 10-step score. For the observers these are vascularity, pigmentation, pliability, thickness, surface and relief. For the patients the six items are pain, itching, color, pliability, thickness and relief. The lowest total score is six and reflects with normal skin, whereas the highest score 60 reflects the worst imaginable scar. The second number of the observer and patient score represents the overall impression of the scar in a 10-step scale, in which ten is the worst imaginable scar. Because re-pigmentation is an important item in our study, this item is listed separately. Again a 10-step scale is used in which 1-4 is mild re-pigmentation, 5-7 moderate, 8-10 severe re-pigmentation. Age s (month) is age at first out patient clinic. Age f (yr) is the age at time of follow-up.

<table>
<thead>
<tr>
<th>o.</th>
<th>sex</th>
<th>age (mth)</th>
<th>age (yr)</th>
<th>size GCMN</th>
<th>location GCMN</th>
<th>POSAS observer</th>
<th>POSAS patient</th>
<th>Pigmentation</th>
<th>Cosmetic outcome</th>
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<tbody>
<tr>
<td>1.</td>
<td>f</td>
<td>12</td>
<td>14</td>
<td>3%</td>
<td>parietal and occipital</td>
<td>14 / 2</td>
<td>26 / 5</td>
<td>none</td>
<td>good</td>
</tr>
<tr>
<td>2.</td>
<td>m</td>
<td>8</td>
<td>12</td>
<td>2%</td>
<td>frontal</td>
<td></td>
<td>none</td>
<td>good</td>
<td></td>
</tr>
<tr>
<td>3.</td>
<td>f</td>
<td>16</td>
<td>18</td>
<td>3%</td>
<td>frontal</td>
<td>24 / 7</td>
<td>35 / 6</td>
<td>mild</td>
<td>brow elevation</td>
</tr>
<tr>
<td>4.</td>
<td>f</td>
<td>4</td>
<td>11</td>
<td>2%</td>
<td>zygoma and temporal</td>
<td>20 / 6</td>
<td>37 / 4</td>
<td>none</td>
<td>scar widened</td>
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<td>5.</td>
<td>m</td>
<td>8</td>
<td>12</td>
<td>4%</td>
<td>frontal and temporal</td>
<td>9 / 2</td>
<td>18 / 7</td>
<td>none</td>
<td>good</td>
</tr>
<tr>
<td>6.</td>
<td>m</td>
<td>4</td>
<td>10</td>
<td>5%</td>
<td>occipital</td>
<td>7 / 3</td>
<td>6 / 1</td>
<td>none</td>
<td>good</td>
</tr>
<tr>
<td>7.</td>
<td>f</td>
<td>4</td>
<td>5</td>
<td>2%</td>
<td>temporal</td>
<td>11 / 4</td>
<td>24 / 4</td>
<td>none</td>
<td>good</td>
</tr>
<tr>
<td>8.</td>
<td>f</td>
<td>9</td>
<td>8</td>
<td>2%</td>
<td>temporal and frontal</td>
<td>25 / 5</td>
<td>29 / 6</td>
<td>mild</td>
<td>brow elevation</td>
</tr>
<tr>
<td>9.</td>
<td>f</td>
<td>5</td>
<td>7</td>
<td>4%</td>
<td>parietal and occipital</td>
<td>13 / 3</td>
<td>37 / 8</td>
<td>none</td>
<td>good</td>
</tr>
<tr>
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<td>3%</td>
<td>frontal and temporal</td>
<td>18 / 4</td>
<td>10 / 1</td>
<td>none</td>
<td>good</td>
</tr>
<tr>
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<td>6</td>
<td>6</td>
<td>2%</td>
<td>parietal</td>
<td>15 / 1</td>
<td>15 / 1</td>
<td>none</td>
<td>good</td>
</tr>
<tr>
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<td>f</td>
<td>4</td>
<td>6</td>
<td>3%</td>
<td>temporal</td>
<td>14 / 3</td>
<td>6 / 1</td>
<td>none</td>
<td>brow ptosis</td>
</tr>
<tr>
<td>13.</td>
<td>f</td>
<td>4</td>
<td>6</td>
<td>3%</td>
<td>occipital</td>
<td>9 / 2</td>
<td>7 / 1</td>
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<td>good</td>
</tr>
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<td>13</td>
<td>5%</td>
<td>tempoparietal and frontal</td>
<td>14 / 3</td>
<td>24 / 4</td>
<td>mild</td>
<td>brow ptosis</td>
</tr>
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<td>m</td>
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<td>5%</td>
<td>temporal</td>
<td>23 / 7</td>
<td>20 / 7</td>
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<tr>
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<td>m</td>
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<td>3</td>
<td>3%</td>
<td>temporal</td>
<td>14 / 3</td>
<td>16 / 5</td>
<td>none</td>
<td>good</td>
</tr>
<tr>
<td>17.</td>
<td>m</td>
<td>4</td>
<td>3</td>
<td>5%</td>
<td>parietal and occipital</td>
<td>21 / 4</td>
<td>21 / 7</td>
<td>none</td>
<td>good</td>
</tr>
</tbody>
</table>

of the patients. Both patients and observers scored the scars as moderate (table 1). In addition, parents scored the over all impression of the scar formation in the POSAS scale as moderate (4-6), which is comparable with the overall impression of the observer scores. In average the scar width was four to five millimetres.

In seven patients (18% of the tissue expanders) eight expander related complications developed: exposure (four), wound infection (two), and implant leakage (two). In one patient two expander related complications developed, namely infection and exposure. In all cases the expander was removed and a new expander was placed. In five patients
reconstruction related complications were seen; alopecia (one), brow elevation (two) and browptosis (two). Patient shown in figure 2 has a minor browptosis. Occasionally a minor “bathtub“ depression was seen at the scalp, immediately after TE, due to pressure of the expander on the skull. This depression was left untouched and was not noticed at the time of evaluation.

In only three patients mild re-pigmentation was seen, which is comparable with a score two to four on a 10-step scale. Special attention was paid to the patterns of hair growth. In line with our expectations, the GCMN located at the occipital and parietal region of the scalp showed the best cosmetic outcomes and had no disturbance at all of hair growth. None of the patients developed a malignant melanoma during follow-up.

Some parents remembered the expansion period as a psychological war of attrition, but all parents were pleased with the cosmetic results. Moreover, they were pleased that the surgical procedures took place at such a young age; none of the children recalled the reconstruction period. No absence at school or psychological problems have been seen in these children.

**Discussion**

In this paper we show that GCMN at scalp or forehead can be excised completely after early tissue expansion. By means of a validated evaluation tool we showed good cosmetic results. In our study period, covering nine years, no cases of malignancy were observed. Due to the small amount of patients, statistical analyses could not be performed.

In seven patients (18% of the tissue expanders) expander related complications occurred; exposure of the expander, implant failure or infection which all interrupted the reconstruction program. Our complication rate is comparable with the figures of 9 to 24% reported in previous publications. 7,11,12,13

In five patients minor (reconstruction) complications developed of whom two had a minor elevation of the brow. Although seemingly obvious, GCMN located at a temporal or frontal region are prone for disturbing the symmetry of the brow position.

When the pivot-point of the expanded rotation flap had a dog ear on the medial side of the brow, we did not want to extend the scar between the eyebrows and the dog ear was left to shrink in time. If insufficient shrinkage took place, a small correction was planned. The amount of expansion differs for each patient and therefore their cosmetic outcome.

Bauer and colleagues highlighted the benefits of expanded transpositions flaps for treatment of GCMN or complex defects in each body region, based on two decades of experience. 8 Initially Bauer used tissue expander to reconstruct the alopecia secondary to
previous grafting or to narrow widened scars in the early patients who had initial grafting followed by hair baring flaps. Nowadays Bauer routinely places tissue expanders prior to excision of the naevus. We used in all of our patients the same technique and always placed the tissue expander prior to excision of the GCMN. We agree with Bauer that this technique should be the treatment modality of care for GCMN at the scalp. We must on the other hand, also keep in mind that serial excision is a good alternative treatment of the smaller CMN of the scalp region, especially in young infants. Also, tissue expansion in children is still known to have a higher complication rate compared to adults and therefore an alternative approach could be close follow up until the age tissue expansion is tolerated.

In our study re-pigmentation did almost not occur and implicates the accurate and complete removal of the GCMN. We feel that any other incomplete removal technique, such as laser therapy, at the scalp should not be performed. Ostertag reported 7 patients with GCMN at the scalp treated with Erbium-Yag laser. Six patients developed little spots of re-pigmentation and in one patient 80% of pigmentation returned within three months. The potential of laser therapy has been justified on the basis of the superficial position of the majority of the pigment in GCMN. One should keep in mind that deeper naevus cells remain when using this technique.

Seldomly, GCMN located on the scalp, but mainly in the midline of the back can develop neurocutaneous melanosis, which has a rapid progressive, fatal prognosis. Frieden and colleagues advocate to make MRI scan of all patients with GCMN at the scalp or back, in order to detect neurocutaneous melanosis. We feel that in neurological symptomatic patients MRI scan is absolutely indicated; an abnormal scan confirms the worse prognosis and indicates a conservative treatment and the patient should be observed closely by a neurologist. In asymptomatic patients it is debatable if MRI scan is indicated; the neonates must have general anaesthesia in order to perform the MRI scan and outcome does not influence the choice of treatment. In three patients we have made a MRI scan with a negative outcome. None of our patients show any kind of neurological symptoms or developed a melanoma within an average follow-up of 8.7 years.

In summary tissue expansion is a good method for removing GCMN located at the scalp or forehead with a good cosmetic end-result. Performing tissue expansion at a young age is advisable.

Acknowledgements

The authors like to thank PPM van Zuijlen for providing the POSAS scale.
Figure 1a. Giant CMN covering mostly occipital region of the scalp of a one year old girl (patient 1 in Table 1)

Figure 1b. Two tissue expansion (8x5 cm and 6x10cm) were placed in a subgaleal plane

Figure 1c. Results after 6 months follow-up period
Figure 1d. Results after 12 years of follow-up

Figure 2a. Giant CMN covering half of the forehead and temporal region of a 8 months old boy (patient 5 in Table 1)
Figure 2b. Two tissue expanders (16x 10cm and 8x6 cm) are placed at a subgaleal plane

Figure 2c. Results after a follow-up of 8 months

Figure 2d. Results after a follow-up period of 10 years. Note the minor brow ptosis
References


